Surgical Correction of Transposition of the Great Vessels: * A Successful Complete Correction

Lt. Col. Elmore M. Aronstam, MC, U.S.A., Col. Thomas H. Hewlett, MC, U.S.A., Col. James A. Orbison, MC, U.S.A., Lt. Col. Robert B. Franklin, MC, U.S.A., Maj. Leon M. Dixon, MC, U.S.A.

From Cardiovascular Surgery Service, Department of Medicine, and Cardiology Service, Fitzsimons General Hospital, Denver, Colorado

DESPITE improved diagnostic ⁸ and surgical technics, transposition of the great vessels remains a most difficult lesion to correct. We have found reports of only six complete corrections with survival ^{3, 6, 9} in the American literature. For this reason our experience may have value in future treatment of this anomaly and is reported herewith.

Case Report

This 18-pound boy was 16 months old at the time of total corrective operation. A normal birth (8 pounds, 2 ounces) had followed an uncomplicated pregnancy. He was cyanotic at birth. His course was marked by poor weight gain and intensification of cyanosis with feeding and crying. Though he became fatigued easily there was no syncope, hypertension, paresis, or squatting. Upper respiratory infections recurred frequently. At nine months of age he neither walked nor crawled. Cardiac catheterization and angiocardiogram performed at nine months showed a complete transposition of the great vessels with intact interventricular septum. The compensating defect allowing survival was an atrial septal defect. At ten months of age a closed surgical procedure was performed at which a high atrial septal defect was created and a branch of the distal right pulmonary artery was anastomosed to the proximal stump of the azygos vein. The child improved following this procedure, becoming less cyanotic and more active. At 13 months of age a month-long series of upper respiratory infections followed by severe pneumonia weakened the child markedly. It was decided to get the child in the best possible physical condition and proceed with total venous correction, since he would not tolerate further respiratory infection.

On May 24, 1962, through a sternal splitting incision, operation was performed using a roller pump, heat exchanger and disc oxygenator set up to infant's proportions. The internal mammary artery and the inferior vena cava were used for recording pressure. The lesion was an obvious transposition. The anastomosis made at the previous operation was intact. A left superior vena cava joined the right close to its entrance in the heart. The left internal iliac artery approached by extraperitoneal incision was used for arterial catheterization. Venous catheters were inserted in both venae cavae through a single purse string suture through the right atrial appendage. As the pump started the child was cooled to an eventual low point of 16° C. The pump was then turned off for 19 minutes. Rewarming was initiated after this period, using the left atrial appendage for venous drainage. Heparin was administered in dosage of 2.0 mg./Kg. as was Polybrene for later neutralization.

As the patient was cooling, a vertical, anterolateral, right atriotomy and a smaller vertical incision parallel to this over the insertion of the right pulmonary veins in the left atrium, were made. The operative high atrial septal defect was seen, as well as a smaller inferior congenital one. The anterior rim of the high atrial septal defect was sutured to the anterior edge of the pulmonary vein incision obliterating this defect. The atrial septum was incised in such a fashion that it could be sutured to the posterior portion of the left atrium, thus excluding the pulmonary veins from the left atrium. At this point the child's temperature was 16° C. and the pumps were turned off. The venous cannulae were withdrawn and the posterior edge of the right atriotomy was sutured

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to the anterior margin created by incision of the atrial septum. This directed caval blood into the left atrium. The anterior margin of the right atriotomy incision was sutured to the posterior edge of the incision of the right pulmonary vein insertion. Thus pulmonary venous blood was directed to the right atrium. The patient was rewarmed using the left atrial appendage for venous return. Pump time was one hour and thirty-three minutes. In effect, a Senning type 9 repair was performed.

Severe metabolic acidosis developed on the evening of operation. This was successfully treated with Tris (hydroxymethyl) aminomethane. Unexplained fever persisted during the first two post-operative weeks. Careful attention to digitalis dosage and fluid administration was necessary as intermittently there was some liver enlargement attributed to heart failure. At two weeks the patient developed a left chylothorax, though his left chest had never been entered. Repeated aspiration and finally insertion of a chest tube resulted in disapperaance and a clear x-ray.

The patient eventually recovered but is still maintained on digitalis. Preoperative arterial oxygen saturations recorded at 40 and 41 per cent have changed to postoperative values of 91 and 92 per cent. He is not cyanotic. He has slowly gained strength and begun to use extremities on which he never walked or crawled before in his young life.

Discussion

Earliest fruitful attempts at palliation of this defect in the United States were those of Blalock and Hanlon.^{4, 5} By creation of a high atrial septal defect better mixing of oxygenated and desaturated blood was obtained. Partial improvement could be obtained in patients with amenable variants of transposition.

Moss,⁷ in 1961, reported a maneuver, ancillary to that of Blalock and Hanlon, anastomosing the cardiac end of a branch of the right pulmonary artery to the proximal stump of the azygos vein. This is an added palliative technic.

Baffes' procedure ² shunting the inferior vena cava to the left atrium and directing the right pulmonary veins to the right atrium resulted in favorable results in 70 per cent of his patients. This operation is

done only if pressure in the pulmonary artery is over 200 mm. of water. For lower pressures he uses other shunt procedures, as the pulmonary artery is not well perfused below this pressure.

Based somewhat on the material of Albert,¹ in 1959, Senning ⁹ described a method of total venous correction of transpositions by intra-atrial rearrangement; systemic venous return was directed to the left atrium and pulmonary venous return to the right atrium. The procedure necessarily must be done on bypass with deep hypothermia. Kirklin ⁶ reported four of 11 survivors using a repair based on the Senning operation. A recent successful case was reported by Barnard and his associates ³ from South Africa. A modification of the Senning operation was used.

Obviously a seriously ill, cyanotic infant is a poor candidate for any operative procedure, particularly an open-heart procedure. However, we are only beginning to appreciate how effectively small children and infants can be handled on bypass and in the postoperative period. As system and technics are bettered it is to be suspected that total correction occasionally can be done in those below a year of age. Of importance is the adequate diagnostic study of these infants in order that the surgeon may know exactly what pathological and physiological problem must be solved at the open operation. Noonan and his colleagues 8 certainly make this clear in their excellent collective review on transposition of the great vessels. Concomitant coarctation, pulmonary stenosis, VSD, PDA and atrial septal defects, complicates the procedure and requires careful preoperative planning.

Palliative procedures must be employed until we can consistently demonstrate success with open procedures in these younger children. The use of a Blalock-Hanlon, Moss or Baffes type procedure obviously does not preclude later total correction.

Summary

A successful open operation for transposition of the great vessels has been reported in a 16-month old infant. Brief discussion of transpositions has been pursued.

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Book Review

Schiff, L.: Diseases of the Liver, Second Edition, Philadelphia, J. B. Lippincott, 1963, \$25.00

With our increasing awareness of the importance of liver function in health and disease, Leon Schiff and the contributing authors have done an excellent job in presenting in a clear and easily readable form the very significant aspects of the anatomy, physiology and pathology of the liver.

The well-written chapter on liver disease in infancy and childhood will be of great interest to the pediatrician. The technic, indications and general appraisal of changes in hemodynamics in cirrhosis and noncirrhotic lesions

with portal hypertensions are dealt with extensively in the chapters on portal venography and monometry. Surgical procedures on the common duct, together with operative diagrams will surely continue to be of great interest to the surgeon. The controversial topic of the so-called complete liver profile tests is very well handled by the authors. The many diagrams, photomicrographs and x-rays, original ideas combined with abundant references makes this one of the most complete texts on the liver and consequently should be well accepted by most clinicians as well as pathologists.

Its simple but dignified binding and fine quality paper are a real credit to the publisher.—Domenic F. Coletta, M.D.